

Congenital dislocation of the patella

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Abstract

Purpose Congenital dislocation of the patella is permanent and manually irreducible, and it manifests immediately after birth with flexion contracture of the knee, genu valgus, external tibial torsion and foot deformity. We retrospectively reviewed the results of operative treatment of seven knees in six patients with congenital dislocation of the patella.

Methods The age of the six patients at diagnosis ranged from 8 days to 3.6 years, with an average of 1.3 years, and their age at the time of operation ranged from 0.6 to 3.9 years, with an average of 2.1 years. Serial casting and/or a brace was attempted before surgery in five of seven knees, leading to improvement in the flexion contracture of the knee. All knees were treated operatively in combination with lateral release, medial plication, V-Y lengthening of the quadriceps, medial transfer of the lateral patellar tendon and posterior release of the knee.

Results Although these deformities were noticed at birth in all seven knees, diagnosis was delayed in three knees due to the low suspicion of the disease and invisible patellae on radiographs. Ultrasonography confirmed the diagnosis of dislocation. The patella was centered in the groove of the femoral condyle after surgery in all knees, but subluxation of the knee with flexion was observed in one knee in which the operation was performed at 3.9 years. Genu valgus and external tibial torsion improved after surgery in all knees. The operated knee was mobile in

all cases, with less than 10° flexion contracture of the knee. Flexion contracture did not increase in any of the knees.

Conclusion Congenital dislocation of the patella should be suspected in every patient with knee flexion contracture, genu valgus, external tibial torsion, foot deformity and delayed walking. Successful results were obtained when the operation was performed in younger children. Other procedures, such as the semitendinosus tenodesis or tendon transfer, might have to be combined to achieve better stability with flexion in older children.

Keywords Congenital dislocation · Patella · Quadriceps malrotation · Operative treatment

Introduction

Congenital dislocation is permanent and irreducible, and it manifests immediately after birth with flexion contracture of the knee, genu valgum and external tibial torsion. It should be distinguished from patellar instability, as well as from recurrent, habitual or permanent reducible dislocations [1–3]. Previous studies [4–7] have discussed patellar dislocation as a congenital dislocation associated with Down syndrome and nail-patella syndrome, but the onset of the former occurs after birth and should be distinguished from the latter [2, 3]. If these criteria are used, congenital dislocation is rare.

We treated this condition using a combination of lateral release, medial plication, V-Y lengthening of the quadriceps, medial transfer of the lateral patellar tendon and posterior release of the knee. The purpose of the present study was to review and evaluate our experiences in the operative treatment of knees with congenital dislocation of the patella.

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Patients and methods

Between 1980 and 2005, seven knees in six patients (two boys and four girls) were operatively treated at our institution for congenital dislocation of the patella. The details of the six patients are shown in Table 1. There were five unilateral and one bilateral patella dislocations. The age of the patients at diagnosis ranged from 8 days to 3.6 years, with an average of 1.3 years, and their age at the time of operation ranged from 0.6 to 3.9 years, with an average of 2.1 years. Patellar dislocations associated with Down syndrome, nail-patella syndrome, Kabuki syndrome or Ellis-Van Creveld syndrome were excluded in the present study.

Patellar dislocation was permanent and irreducible in all knees. Flexion contracture of the knee, genu valgus, external tibial torsion and foot deformity were present in all knees. Ultrasonography and MRI were performed to obtain a diagnosis and preoperative evaluation in all patients. Preoperative flexion contracture of the knee ranged from 5° to 60°, with an average of 28°. Associated calcaneovalgus foot deformity was present in four feet, congenital vertical talus was observed in two feet, and talipes equinovarus deformity in one foot. Three of six patients (cases 4, 5 and 6) who were operatively treated after walking had a severe limp before surgery.

Treatment

Serial casting and/or a brace was attempted in five of seven knees, leading to improvement in the flexion contracture of the knee. Serial casting and/or a brace was not used in one patient whose cardiovascular complications had been treated in another department. All knees were treated operatively in combination with lateral release, medial plication, V-Y lengthening of the quadriceps, medial transfer of the lateral patellar tendon and posterior release of the knee.

Operative technique

Under general anesthesia, the patient was placed in the supine position. A curved incision was made laterally starting from the distal third of the femur, extending superomedially to the patella, and ending medially to the tibial tuberosity. Lateral release was performed by dividing the quadriceps from the fibrous adhesions to the iliotibial band and the lateral intermuscular septum. The lateral capsule was incised laterally to the dislocated patella and along the lateral border of the patellar tendon to the tibial tuberosity. The medial capsule and retinaculum were incised medially to the patella. The patella was released from the lateral aspect of the lateral femoral condyle and

Table 1 Patient data

Case	Sex	Side	Underlying disease	Age at diagnosis	Treatment before operation	Knee flexion contracture (°) before operation	Age (years) at operation	Operative treatment	Complications	Duration (years) of postoperative follow-up	Knee flexion contracture (°) at follow-up
1	F	L	AMC	8 days	Serial casting, brace	10	0.6	LR(PS), MP, QL, LPT		6.9	10
2	F	L	Unknown	9 days	Serial casting, brace	35	0.6	LR(PS), MP, QL, LPT		2.0	0
3	M	R	Multiple congenital anomalies	8 days	None	60	1.7	LR(FG), MP, QL, PR		2.3	10
		L		8 days	None	60	1.8	LR(FG), MP, QL, PR		2.3	10
4	F	L	Chondrodysplasia punctata	2.6 years	Brace	5	2.8	LR(FG), MP		4.1	5
5	M	R	Congenital fibular deficiency	2.9 years	Brace	10	3.2	LR(PS), MP		10.2	10
6	F	R	Unknown	3.6 years	Serial casting, brace	15	3.9	LR(FG), MP, QL, LPT	Subluxation with flexion	5.7	0

AMC arthrogryposis multiplex congenita, LR lateral release, PS patch with a polytetrafluoroethylene (Gore-Tex) sheet, MP medial plication, QL V-Y lengthening of the quadriceps, LPT medial transfer of the lateral patellar tendon, FG patch with a free graft taken from the abundant medial capsule and retinaculum, PR posterior release of the knee

reduced into the typically underdeveloped shallow groove of the femoral condyle.

The tracking of the patella was checked carefully. In the five knees in which knee flexion was restricted due to the shortened quadriceps, the quadriceps tendon was lengthened proximal to the patella by V-Y plasty. In three of these five knees, in which mild lateral subluxation remained, the lateral half of the patellar tendon was carefully detached at its insertion from the cartilaginous tibial tuberosity and was sutured medially at the midline of the upper tibia. The lateral subluxation improved as a result of the lateral half of the patellar tendon transfer in all five knees. In the two knees with severe knee flexion contracture, posterior release was performed using two vertical incisions in the prone position after the anterior part of the surgery. Full extension was achieved by Z- or fractional lengthening of the hamstrings and semi-membrane and semitendinosus tendons; posterior capsular release was not performed.

The lateral capsular defect was patched to prevent synovial fluid leakage in all knees. A free graft taken from the abundant medial capsule and retinaculum was used in four knees and a polytetrafluoroethylene (Gore-Tex) sheet (W.L. Gore and Associates, Flagstaff, AZ) was used in three knees. Medial plication was performed in all knees. The medial capsule and retinaculum with a part of the vastus medialis were drawn laterally and sutured antero-laterally to the patella.

A long-leg plaster cast was applied to hold the knee in slight flexion for 2–3 weeks after surgery. Active and passive exercises of the knee were initiated after cast removal.

Results

The follow-up period ranged from 2.0 to 10.2 years, with an average of 4.8 years. Three of seven knees were not accurately diagnosed with patellar dislocation at our initial consultation. Ultrasonography was useful in all patients for evaluating the location of the non-ossified patellae and confirmed the diagnosis of dislocation. MR imaging was useful for preoperative evaluation and clearly visualized the malrotated quadriceps muscle with insertion of the patella tendon, the hypoplastic dislocated patella and the undeveloped intercondylar groove of the femoral condyle. Serial casting and/or braces were attempted before surgery in five of seven knees and were effective in reducing flexion contracture.

At the time of writing, the reduced patella was centered in the groove of the femoral condyle in all knees, but lateral subluxation with flexion persisted in one knee; the patient complained of femoropatellar pain after walking for long

distances. Genu valgus and external tibial torsion improved after surgery in all knees. The operated knee was mobile in all cases, with less than 10° flexion contracture of the knee. Flexion contracture did not increase in any of the knees. The average flexion contracture of the knees was improved to 6° (0°–10°) with an average improvement of 22° (0°–50°). The preoperative limp in the three patients (cases 4, 5 and 6) improved after surgery. Two patients without the underlying disease (cases 2 and 6) had no limp at the time of writing.

Discussion

There has been great confusion regarding the classification of patellar dislocation in children [2, 3]. Many previous reports include patellar dislocations found in Down syndrome or nail-patella syndrome in the group of congenital dislocations of the patella [4–7]. The patellar dislocations associated with Down syndrome, nail-patella syndrome, Kabuki syndrome, and Ellis-Van Creveld syndrome are considered to occur after birth, during childhood or adolescence, and to gradually become permanent and irreducible; they were excluded from the present study. This condition was observed in children with joint laxity and occurs more or less late during growth, and much more frequently in teenagers [1]. The term developmental dysplasia of the patella (DDP) accurately reflects the variable characteristics of this condition [1].

Congenital dislocation of the patella is a different entity and should be distinguished from other patellar dislocations [1–3]. It is characterized by a relative stiffness, a short quadriceps, and a major femoropatellar dysplasia. Congenital dislocation is permanent and irreducible, and manifests immediately after birth with flexion contracture of the knee, genu valgum and external tibial torsion (Fig. 1a).

Although these deformities were noticed at birth in all seven knees, diagnosis was delayed in three knees due to the low suspicion of the disease and, because the patella ossifies between 3 and 5 years of age, the invisibility of patellae on radiographs (Fig. 1b). When flexion contracture is severe, active extension of the knee is impossible because the quadriceps mechanism works as a knee flexor. In patients whose flexion contracture is mild or is improved by serial casting and/or a brace, active extension of the knee becomes possible with mild impairment of walking. Thus, patients with congenital patellar dislocation are often misdiagnosed with a neuromuscular disease or muscular dystrophy. Congenital dislocation of the patella should be suspected in every patient with arthrogryposis, skeletal dysplasia or other related syndromes, especially if the patient presents with knee flexion contracture, genu valgus,

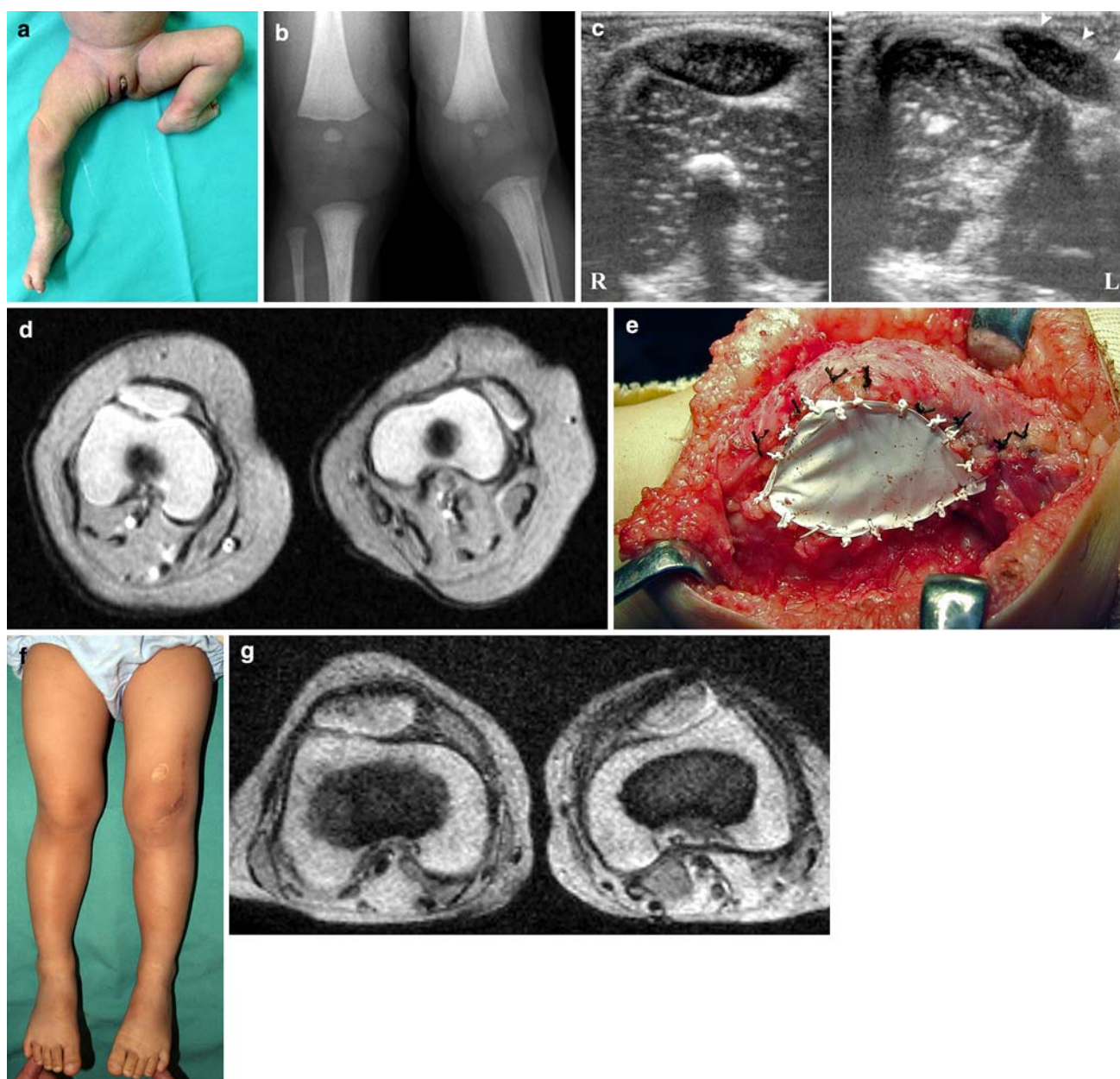


Fig. 1 **a** A 9-day-old girl (case 2) presented with severe knee flexion contracture and external tibial torsion. Active extension of the knee was impossible. **b** Anteroposterior radiograph at 9 days showing left genu valgus and external tibial torsion. **c** Axial ultrasonography shows hypoplastic, laterally dislocated patella (*arrowheads*). **d** MR imaging at 0.5 years showing a laterally dislocated patella adherent to the lateral condyle and an underdeveloped, shallow groove of the

femoral condyle. **e** Surgery at 0.6 years consisting of lateral release, medial plication and V-Y lengthening of the quadriceps tendon. The lateral capsular defect was patched with a polytetrafluoroethylene sheet. **f** Clinical photograph at 2.6 years showing no knee flexion contracture nor external tibial torsion. **g** MR imaging at 2.6 years showing the reduced patella in the groove of the femoral condyle

external tibial torsion, foot deformity and delayed walking [4]. A careful physical examination is necessary, focusing on palpation of the patella, which is small, impossible to find at the anterior aspect of the knee, relatively fixed and lies laterally on the lateral condyle. Ultrasonography is easy to perform and is useful for locating the patella and confirming the diagnosis (Fig. 1c) [3, 8–11]. MR imaging allows visualization of fine anatomic details and

relationships between the involved structures of the extensor mechanism and should be performed as a part of preoperative assessment and planning (Fig. 1d) [10, 11].

The only possible treatment for congenital dislocation of the patella is surgical. Serial casting and a brace are effective in reducing flexion contracture of the affected knee, but genu valgum, external tibial torsion and subluxation of the tibia inevitably develop without surgical

intervention. Stanisavljevic et al. [12] reported successful results of the extensive lateral release and anterior transposition of the entire quadriceps mechanism based on their suggested pathogenesis of malrotation of the quadriceps myotome during embryonic life. However, the entire release of the quadriceps requires a wide incision and extensive lateral dissection. The present operative technique of lateral release was less extensive than the Stanisavljevic procedure [12]. We did not extend the incision to the level of the greater trochanter and did not strip the quadriceps extraperiosteally. Our incision was limited to the mid-thigh, and our lateral release was performed by dividing the quadriceps from the fibrous adhesions to the iliotibial band and lateral intermuscular septum. V-Y lengthening of the quadriceps, medial transfer of the lateral patellar tendon and posterior release of the knee were combined according to the severity of the deformity. Bensahel et al. [1] and Eilert [9] also describe the effectiveness and good outcome of a similar procedure using a combination of lateral release, medial plication, lengthening of the quadriceps, and medial transfer of the patellar tendon.

The operative findings in congenital dislocation of the patella have been described as follows [1–9, 12]: (1) malrotation of the quadriceps muscle with insertion of the patellar tendon on the anterolateral tibia, (2) contracture of the quadriceps and iliotibial band, (3) a hypoplastic patella with a flattened articular surface fixed on the lateral knee, (4) an undeveloped, shallow anterior intercondylar groove of the femoral condyle, (5) contracture of the lateral capsule and thickening of the medial capsule, (6) genu valgus and external tibial torsion and (7) lateral subluxation of the tibia. These were consistent with the findings in the present series.

We used the polytetrafluoroethylene (Gore-Tex) sheet to make patches for the large lateral capsular defect to prevent synovial fluid leakage and adhesions to the quadriceps muscle and the surrounding tissues (Fig. 1e). The sheet has little reaction or adhesion to living tissue and has been used for dural repair, artificial vessels and patches for pericardium. To our knowledge, there is no orthopedic literature on the usage of the sheet except for our case report [13] in which the sheet was used in the ankle joint. Long-term follow-up is necessary to fully clarify the efficiency and safety of the sheet.

The operation should be performed early, as soon as the diagnosis is confirmed, and preferably before 1 year of age [3]. In our two knees, which were operatively treated before 1 year of age, early operation made it possible to reconstruct almost normal knee function and to provide the

patient with a normal gait without delay at the onset of walking (Fig. 1f, g). Neglected cases require more extensive excisions of contractures to ensure reduction of the patellar dislocation and more complicated surgical reconstructions to ensure patellar stability [3]. Subluxation of the knee with flexion recurred in the knee in which the operation was performed at 3.9 years. Other procedures, such as the semitendinosus tenodesis as described by Galeazzi [14] or tendon transfer as described by Uezaki [15], might have to be combined additionally to achieve better stability with flexion in older children.

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